
THE
Journal
OF
Nervous and Mental Disease

Original Articles.

RIGIDITY OF THE SPINAL COLUMN.*

BY PHILIP ZENNER, A.M., M.D.,

CINCINNATI.

In 1893 Bechterew¹ reported three cases of rigidity and curvature of the spine with peculiar nervous manifestations, considering them to present a special form of disease. Whilst no attention had hitherto been paid to this subject in neurological literature, a number of other publications was called forth by that of Bechterew, the cases being alike, at least, in the presence of rigidity of the spine, though how far they all belong in the same category of disease is doubtful. As the matter has not, so far as I know, received any consideration in American neurological literature, I wish to give a brief review of published cases, as well as to report a few coming under my own observation.

Bechterew² in a second article with the report of additional cases, gives the following résumé of the symptoms presented:

Complete, or nearly complete immobility of a part of, or the entire spin.

A large rounded posterior curvature of the spine, chiefly in the upper dorsal region. The head appeared to be pushed forward or sunken while the part of the spine below the pos-

* Read by title at the twenty-fifth annual meeting of the American Neurological Association, June 14 and 15, 1899.

¹ "Steifigkeit der Wirbelsäule und ihre Verkrümmung als besondere Erkrankungsform." *Neurol. Centbl.*, 1893, p. 426.

² "Von der Verwachsung oder Steifigkeit der Wirbelsäule." *Deutsche Zeitsch. f. Nervenheilkunde* Bd., xi., p. 327.

terior curvature was straight, the normal concavity in the lumbar region being absent.

A series of symptoms that may be looked upon as spinal root-symptoms, present to a varying extent in different cases, such as paresthesia and pains in the extremities, neck, trunk and spine; in a few cases, cramp-like contraction especially in the extremities; impaired sensation in areas supplied by the lower cervical, dorsal, or lumbar nerves; weakness of the muscles of the neck, trunk and extremities; and usually, slight atrophy of scapular muscles; usually little or no pain produced by either percussion, or attempted movement, of the spine.

The electrical reaction of the spinal muscles was sometimes lessened quantitatively, but not otherwise affected. There was some flattening of the chest, and, mostly, abdominal breathing. The joints of the extremities were only occasionally affected. The disease was in all instances slowly progressive, and, while offering no hope for improvement, did not appear to have any direct fatal tendency. His patients were all fifty years old or more, excepting one who was about thirty-five. Heredity or trauma appeared to be the cause in some instances.

Bechterew, while knowing of no autopsies, expressed the opinion that there was progressive ankylosis of the spine, involvement of the spinal roots, and extension of the inflammatory process to the outer layer of the dura and the surrounding connective tissue.

The series of cases reported since Bechterew's have mostly another very pronounced condition, ankylosis of the hips. In some there was disease also of other joints. In his second article Bechterew stated that the only other report similar to his own with which he was acquainted was that in Oppenheim's text-book, under the heading arthritis deformans of the spine. According to the latter the disease is attended by pain in the spine, intensified by effort to move, and spinal root-symptoms attributed to slow compression through growth of bone into the vertebral canal such as intercostal, brachial, and crural pains, and atrophic paralyses. The disease runs a chronic course, with remissions and exacerbations. The diagnosis is based on arthritis in the joints, rigidity of a large part of, or the entire spine, which does not disappear under chloroform, and the spinal root-symptoms.

In the same journal an article of Strümpell's³ appeared in which he gives a clinical picture based upon three cases seen by himself. He describes the disease as remarkable and peculiar, resulting in gradual ankylosis of the spine and hips, without spinal root-symptoms, in fact with almost entire absence of subjective symptoms. One case was examined under chloroform. The spine remained immobile. Rotation of the hip, which was only slightly movable, produced crepitation. The spine was abnormally straight, instead of having the large kyphotic curve of Bechterew's cases. The muscles along the spine appeared abnormally hard and atrophied. His patients were between thirty and forty years of age. He says he knows of no autopsy in such cases, but, on account of the crepitation of the hip in one of his cases, believes that there is a chronic inflammatory process affecting the joints of the vertebræ as well as the hip, yet is in doubt whether this is in etiological relation with other arthritic processes.

Baumler⁴ in a later issue of the same journal gives a brief account of a case seen by him in 1873, with his opinion as to the nature of the disease. A man of twenty-three had inflammation of both hip-joints at seventeen; the lower part of the spine becoming affected, perhaps, at a later period. When seen by Baumler, in addition to rigidity of the spine and hips and lessened mobility of the right shoulder, the head was bent forward, there was thickening of the tissues about the cervical spine, and the patient suffered with constant pain in the neck.

Baumler believes that the state of the spine in these cases is due to mechanical causes. He says, when the spine of normal individuals is held long in one fixed position, on account of the occupation or the like, local pain and tenderness are produced which require a long rest for their removal. He thinks that in the kind of cases we are considering, inflammation of the hips or of part of the spine, and, possibly in addition, a strained position due to the special occupation, cause the as yet unaffected spine to be held in a fixed position, with no normal wholesome

³ "Bemerkung über die chronische ankylosirende Entzündung der Wirbelsäule und der Hüftgelenke." *Deutsche Zeitsch. f. Nervenheilkunde* Bd., xi., p. 338.

⁴ "Ueber chronische ankylosirende Entzündung der Wirbelsäule." *Deutsche Zeitsch. f. Nervenheilkunde* Bd., xii., p. 177.

play of the parts. As a result there are firstly, abnormal circulatory conditions, congestion in the immobile parts, and, subsequently, from the presence of micro-organisms, inflammation. In his own case the patient was a writer, and the rigidity of hips and larger part of spine required a constant strained position of the cervical spine—a forcible bending over in order to read his books—and inflammation about the cervical spine ensued. After a long rest the inflammation and pain about the neck disappeared, and the man resumed his work, though the rigidity remained. Baumler supposes that such mechanical influences were the chief features in Bechterew's cases, whilst the same, with an additional inflammatory concomitant, would explain his own and Strümpell's cases. In others, he thinks the condition may be due to inflammation alone.

Marie⁵ reports two cases, and gives as characteristics of the disease, complete rigidity of the spine, with more or less pronounced ankylosis of the hips and shoulders (therefore "rhizomélique"), the small joints remaining free. In his cases the hips were quite ankylosed, there was lessened mobility in shoulders and knees, a pronounced rounded posterior curvature of the upper half of the spine, the lower half straight without the normal concavity in the lumbar region; there were exostoses on the sacrum and cervical spine, flattening of the chest and atrophy of muscles in the dorsal and gluteal regions.

Mutterer⁶ reports a case like Marie's in the condition of the spine, hips and shoulders. The soft parts around the cervical spine were decidedly hard, long muscles of the back flaccid and atrophied, and gluteal muscles atrophied. The knees were swollen, there was no tenderness over the vertebrae, and nothing abnormal, exostoses, etc., to be detected. The patient, a man of fifty-eight, was often kept awake at night by shooting pains in his legs. Otherwise he was in good health. His first symptoms were pains in the loins eight years before. Four years later there was some rigidity, shooting pains in the legs and some difficulty of walking. The rigidity slowly passed from below upwards, the cervical region being last affected, its involvement being accompanied by severe pain.

⁵ "Sur le spondylose rhizomélique." *Rev. de Médecine*, April 10, 1893.

⁶ "Zeitschr. f. Nervenheilkunde Bd., xiv., p. 144.

Popoff⁷ reports a case in a man of twenty-three, distinguished from the other reported cases in its rapid development. The disease was apparently due to the patient's being in the water five hours fishing. The same evening he had fever and pain in the loins and knees. Within two months, during which time there was pain in the spine, there developed the condition of the spine noted in Bechterew's cases, rigidity, large kyphotic curve in its upper half and absence of normal convexity in the lumbar region. There was tenderness over the spine, from the sixth dorsal to the third lumbar vertebra. He continued to have occasional pains in the knees, sides and spine. The breathing was abdominal, the gait weak and tremulous. Though the spine was rigid the head could be moved freely.

This case is unlike the others, excepting Bechterew's, in the freedom of the joints. But during his illness the thumb was swollen and painful for a period of six weeks, and, six years prior to this period, the patient had had inflammation of the wrists for a year, terminating in permanent ankylosis, all pointing to arthritic tendencies.

In how far all of these cases belong to the same category of disease is by no means clear. Bechterew and most of the other authors speak of the absence of post-mortem findings, but Bechterew⁸ has since reported the death from pneumonia of one of his cases, and the post-mortem examination. The very brief report of this examination, under society proceedings, merely states that there was very slight mobility of the spine, that the bodies of the upper dorsal vertebræ were grown together to a larger or smaller extent, that there were osteophytes on the bodies of these vertebræ, and that there were no marked changes in the dura. The posterior roots of some of the dorsal nerves were degenerated. There was a slight grayish discoloration of the root zones of the posterior columns.

We may gain some light on the subject from surgical literature.

Marsh⁹ mentions a case of Fagge in Guy's Hospital in

⁷ "Ueber die Ankylose der Wirbelsäule." *Neurol. Centbl.*, 1899, p. 294.

⁸ *Neurol. Centbl.*, Feb. 1, 1899, p. 143.

⁹ "Rare Forms of Bony Ankylosis." *Brit. Med. Journal*, June, 1895, Vol. 2, p. 1087.

1874—a man thirty-four years of age. A year prior to this time the spine became stiff and formed a rounded curve. The right hip became ankylosed subsequently. The patient died of chest trouble.

At the autopsy there was found ankylosis of the right hip, of the attachments of the ribs to the vertebræ, as well as complete ankylosis of the arches, spinous and articular processes of the dorsal vertebræ. The vertebræ were softened so that they could be cut with a knife. The spine was fragile.

Marsh states that bony ankylosis of the spine is common in osteo-arthritis. Such specimens are to be found in every large pathological museum.

The union results from the direct fusion of component bones, as in Fagge's case, from ossification of the ligaments, or from the formation of bony buttresses connecting the adjoining vertebræ, the joints remaining unaffected. Marsh states, also, that all large museums show specimens of bony ankylosis in extreme lateral curvature, the vertebræ being fused together in the concavity of the curve.

Koehler¹⁰ in an article on an unusual case of spondylitis deformans, describes a case in many respects like the foregoing. A man of sixty-one, six years prior to Koehler's seeing him, noticed some stiffness in the legs, and, soon afterwards, in the spine. The mobility of the knees and hips became gradually lessened, their movement being attended by a creaking sound, and a feeling as if sand were in them. Movement of the head was attended by a sense of cracking in the upper cervical vertebræ. There was some pain on attempted movement, otherwise scarcely any pain. At the end of four years, spine, hips and knees were nearly rigid. At this time he fell on the floor in walking and broke his left thigh in its upper third. He had been in bed six months when first seen by Koehler. At this time he was very emaciated, especially in the muscles of the back, which required a very strong faradic or galvanic current to produce contractions. There was no tenderness over the spine, slight lateral curvature in the dorsal region, but no deformity. The head could be turned or bent to the least possible extent. Otherwise the entire spine from

¹⁰ *Charité Annalen*, 1887, p. 619.

head to sacrum, was found, under chloroform, to be as rigid as a board. On examining by way of the throat there was found to be a number of irregular swellings of the size of beans on the sides of the cervical vertebræ. Osteophytes were also felt upon the sacro-iliac joints. The left hip was totally ankylosed. The right hip and the knees were very slightly mobile, their movement producing crepitation.

Koehler gives the following description of spondylitis deformans. The intervertebral disks fibrillate or are entirely destroyed; sometimes there is distinct growth of new cartilage. Furthermore, supra-cartilaginous exostoses grow on the rims of the vertebral bodies, in part in stalactitic form. They may become united with like growths from the neighboring vertebra, forming a bony clasp which connects the vertebræ together. Rarely is the entire rim of the vertebra involved in this process. Usually only the lateral aspect, or only one side is affected. At the same time there is atrophy of the body of the vertebra, different parts being affected to different degrees, so that it assumes an irregular form as regards height, breadth and depth, and as a result there may be strikingly irregular curvature of the spine. At the same time the ligaments may undergo a bony transformation. This deforming inflammatory process may attack single vertebræ, a number of vertebræ, or even the entire spine.

Koehler quotes Braun as giving the causes of spondylitis deformans in fifty-eight cases—in most of which there was disease of only a few vertebræ—as follows: Syphilis 3, trauma 2, arthritis deformans 3, gout 4, in the rest chronic rheumatism. Koehler believes in his own case the presence of exostoses, and appearance of the joints prove that the disease was arthritis deformans, notwithstanding that this disease almost never leads to ankylosis. The ankylosis of the left hip is due, in his opinion to the long immobility following fracture of the thigh.

Before commenting further on the disease, I will add the report of some cases of my own.

Case 1. M. R., age twenty-nine, merchant. His father, who suffered with diabetes, died of obstruction of the bowel at fifty-eight. His brother and himself are color-blind. No his-

tory of rheumatism in the family. No history of trauma. Excessive venery, no tobacco, nor alcohol.

Nine years ago he had gonorrhea. There was a suspicion at the time of the presence of a urethral chancre, but, so far as he knows he has never had any constitutional symptoms of syphilis. He gives a history of varied pains for a long period of time, even antedating the just mentioned venereal disease. He does not give a very good account or description of these pains, but he has had at times pains in the back, in the neck, the extremities, and, probably is never for a long period altogether free from pain. That in the back seems to have been mostly of the character of lumbago. The pain of which he complains most is of a severe tearing character in the neighborhood of the right loin, coming on in paroxysms, and occurring chiefly at night. Such paroxysms of pain were what especially led him to consult me. He thinks he has observed some stiffness of the back for five years, and that it has tended to increase. He has also had various choreiform symptoms; for eight or ten years, occasional "tic" of eyelids; for a year, occasional "tic" of neck, and also peculiar noises coming from the throat, which are at times made unconsciously, again as a result of a certain sense of constraint. Otherwise he has always enjoyed excellent health.

The examination of the patient apart from the "tics" just spoken of, reveals nothing abnormal, excepting in what relates to the spine. The latter appears to be quite immobile, excepting the upper cervical vertebræ. The head can be rotated freely, and can be bent backwards and forwards, though not quite as much as normally. There is a rounded kyphotic curve in the upper dorsal region, but probably not so large as that depicted by Bechterew. The normal concavity in the lumbar region is absent. When he stands the shoulders are thrown a little backwards, the abdomen and knees a little forwards, the knees slightly flexed. The whole attitude is that of a peculiar bent-over posture. The stiffness of the trunk and all, also gives the gait a peculiar appearance. The muscles of the back respond normally to the electric current, and they do not appear abnormally hard or atrophied. There is no tenderness over the spine. The patient can jump on his heels without producing any pain. He is accustomed to walk long distances with ease. He presents a few symptoms of a spastic character. At times he cannot cross the knees with ease. He cannot fully extend the knee while in a sitting position, apparently on account of strong contractions of the hamstring muscles. Occasionally he feels a sudden contraction of the muscles of the trunk so that for a moment he cannot straighten the body as

well as usual. He always has observed that he is taller in the morning, on rising, than later in the day.

The patient, who came from a southern city, remained under my charge for several months. The treatment consisted mainly in maintaining the supine position for several hours each day, hot applications to the spine during that time, and massage of the muscles. He suffered more or less with pains, mostly of the character of muscular rheumatism, but not severe. He had no recurrence of the paroxysms of severe pains from which he had been suffering. His general health remained good. When he left, the rigidity of the spine, and its appearance remained unchanged.

Case 2. M. S., age forty-four, policeman. A sister has epilepsy. Much addicted to tobacco, not to alcohol. Denies syphilis, but states that he had iritis in 1892. Has been married fifteen years and has healthy children.

In 1884, when his duties were very taxing, and he was subjected to much exposure, he was laid up with lumbago for six weeks. In 1894 "caught cold" and was again laid up with pains in his back and a stiff back. He has never been altogether well since the latter date. His chief complaint has been of pain. This pain is mostly in the loins, chiefly on the left side, and is only felt when making a movement, especially if trying to arise while on his back. He says he awakes with pain several times in the night, possibly the result of turning in his sleep.

As in the first case, apart from what relates to the spine there are no objective symptoms, the muscular strength, cutaneous and muscular sensation, reflexes, special senses, etc., being normal. He presents the same condition of the spine: immobility of all parts excepting the upper cervical region, and a kyphotic curvature in the upper dorsal, and lower cervical region. The head appears pushed forward, though not drooping. The movement of the head is much less than in my first case, there being scarcely any lateral movement, and little flexion or rotation. The kyphotic curve is not marked and the normal lumbar lordosis not altogether lost. The man was originally six feet two and one-half inches in height, now he measures one and three-fourth inches less. There is some tenderness over the vertebræ, especially the lumbar, but greater tenderness over the muscles of the loins. The breathing is distinctly abdominal. Though a tall man the measurements of the chest show only one inch difference between forced inspiration and forced expiration.

This patient was also under observation for several months, with no perceptible change excepting that the tenderness over the spine and muscles disappeared.

Case 3. The third case appears to be altogether different from the other two. The patient is a boy sixteen years of age, brought to me to examine into his mental condition. He had spasms in infancy, and has always been somewhat peculiar in disposition. It is needless for me to enter here into a consideration of his mental condition. Suffice it to say that while he is honest and conscientious, learns easily, has a good memory, etc., he has many mental stigmata of degeneration; is abnormally sensitive, stubborn, given to violent fits of anger, slovenly to the point of indecency, etc. He has also many physical stigmata, thick lips, prominent ears, strabismus, very awkward hands, chest pigeon-breasted, etc. He is also a somnambulist.

He has a large kyphotic curve of the spine, taking in the whole dorsal region. There is, at the same time, no mobility, or very little of the entire spine, excepting in the upper cervical and lumbar regions. On account of the curvature the head is pushed forward, but it is freely movable. There is the normal lumbar lordosis. The breathing is free and normal. There is no tenderness over the spine or back, and he does not suffer, as far as I can learn, has never suffered, with pain in these parts. I could not obtain any satisfactory history of how long this curvature had existed.

The feature common to all the cases mentioned was rigidity of the spine, either a large part of, or the entire vertebral column being involved. In most instances the ankylosis affected also the costo-vertebral joints, indicated by the abdominal breathing. The large kyphotic curve was also a common feature, though absent in Strümpell's, Baumler's, and Koehler's cases. Koehler believed that it always appeared after some time, and was only absent in his own case on account of the patient's long confinement to bed, an explanation which is scarcely satisfactory.

Spinal root-symptoms, spoken of by Oppenheim and Bechterew, were not noted or not pronounced in most instances, and, in general, pain was not very marked.

That the disease was of a distinct arthritic character in some instances seems clearly indicated by the condition of other joints—complete or partial ankylosis, sometimes crepitation—and, in a few cases, by the presence of osteophytes on the vertebræ. It may be that they differed in the character of the arthritis; gouty, rheumatic, arthritis deformans, etc. That all the cases were primarily arthritic is not so clear, not of Bech-

terew's, Popoff's, and my own. Popoff mentioned a case of his own, reported in an earlier publication, in which the curvature of the spine was decidedly reduced under chloroform. The curvature was also lessened by lying a few hours, and it was noticed that the spine was straighter in the morning than in the evening, yet no mobility of the spine could be detected on examining him. In this instance Popoff believed the spinal rigidity was from muscular contractions. In his other case there had been some lessening of the curvature of the spine during the four months the patient was under observation. In this case, in which the whole process was of rapid development, the curvature began to appear within a month of the onset of pain and gradually increased. But it was observed that in the beginning the curvature would disappear when the patient lay upon his back, and only after some months, became permanent.

In my first two cases it impressed me as if the trouble were primarily muscular. The pains were chiefly of the character of muscular rheumatism, there were no joint symptoms, and the first patient spoke of always being taller in the morning when he got up than in the evening, and presented other spastic symptoms.

Mutterer¹¹ quotes Beer, whose report of a like case was inaccessible to me, as saying that the soft parts were the chief seat of the disease.

If muscular contractions be the primary condition in these instances it is not improbable that bony ankylosis finally ensues either like that occurring in lateral curvature, or from a resulting inflammation in the vertebræ or intervertebral disks.

¹¹ Zeitschr. f. Nervenheilkunde, Bd., xiv., p. 144.